SMITH, (R.M.) & PARKER, (A.J.)

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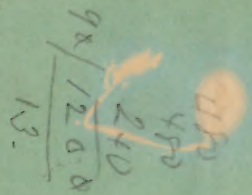

# DISSECTION OF A HUMAN OTOCEPHALIC CYCLOPS MONSTROSITY.

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For description of Case I., see page 9.

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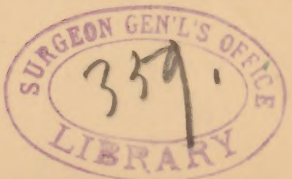
At the request of the Mütter Museum Committee of the College of Physicians of Philadelphia, who obtained possession of the specimen, we were able to dissect the Cyclops monster, described as Case I. in the preceding article.

We find it necessary to make very few additions to the description there given of the external appearances, though dissection enabled us to correct several errors which were unavoidable with the exterior alone for a guide. We find that the monster belongs to the group of *otocephala* of St. Hilaire, and to the division of this group which he calls *edocephala*, characterized by the tendency to fusion of the external auditory apparatus beneath the head, the rudimentary condition of the lower jaw, absence of mouth, and presence of a rudimentary nasal apparatus, in the form of a proboscis, above a single eye placed in the centre of the face.

The specimen referred to us for description was that of a female fœtus, probably, from the undeveloped condition of the larynx and finger-nails, in the eighth month of gestation. The only abnormality was in the development of the head, and our description, therefore, will be confined to that region:—

The general shape of the cranium is normal, with the frontal region at first sight apparently well developed, no suture being perceptible between the frontal bones. The frontal protuberances are represented by two marked prominences at the line of union of the frontal and parietal bones; although, therefore, really malformed, the general appearance of the frontal region is that of a normal skull. The circumference of the head over the frontal protuberances is 26.3 centimetres. The occipital and parietal prominences are well developed, and the occipital and frontal fontanelles occupy their normal positions; the latter, however, being triangular in shape, with the base anterior, instead of quadrangular. Situated 4 cm. behind the frontal fontanelle, and 1 cm. in front of the occipital fontanelle, in the line of the sagittal suture, there is an oval defect in development of the parietal bones, 2 cm. in diameter laterally, and 15 mm. in antero-posterior diameter.



Situated immediately below a quasi glabellum, 15.1 centimetres from the occipital prominence, there is a fleshy, flexible, trumpet-shaped proboscis, 2 centimetres in length, 1 centimetre in diameter at its free end, and 5 millimetres in diameter at its point of attachment. It is covered throughout with integument, continuous with that of the frontal region, which, at its free extremity, is involuted to form a single blind pouch 3 millimetres in depth and in diameter.

Immediately below this rudimentary nasal apparatus, and overhung by it, is situated a single eye, placed in the median line, in a bony orbit of nearly circular outline, the diameter of the palpebral fissure being 16 millimetres. The upper and lower eyelids are feebly developed, and meet at each side in a shallow groove or furrow, extending outwardly 3 millimetres; the upper lid is circular in outline at its margin, and freely separated from the eyeball to the depth of 3 millimetres; eyelashes are well marked, and, although the frontal region is covered with faint downy hair, there is no indication of an eyebrow. The lower eyelid has a crescentic emargination in the median line, where it is attached to the eyeball, the remainder of the lid being free from the eyeball to the depth of 3 millimetres, as in the case of the upper lid. From the feeble development of the eyelids the eyeball is left considerably exposed.

Below the single orbit the superior maxillary bones are easily detected; they are narrow and more flattened laterally than is normal. Below the inferior margin of the superior maxillary bone there is a second fleshy prominence, closely resembling in general outline a normal chin; no signs of an inferior maxillary bone can, however, be detected. At the summit of this tuberosity, which is about 3 centimetres in diameter, and 2 centimetres in height, there is situated a circular opening, 3 millimetres in diameter, communicating with a blind pouch 23 mm. in depth, and of the same apparent breadth.

The neck is somewhat flattened antero-posteriorly. On its upper portion, displaced below the base of the skull towards the median line, are the external auricles, the external auditory meati being represented by two slit-like openings, 23 millimetres apart, occupying their normal relations to the auricles, inclined downward and toward the median line. A probe can be readily passed through these openings into the pharynx. The helix and anti-helix are moderately well-marked, and the anti-tragus and lobe of both ears are present; the tragus is not discernible.

On removing the calvaria it was found that the brain did not completely fill the cranial cavity. The cerebrum was found incompletely divided into two hemispheres, anteriorly no separation having taken place; posteriorly, however, the cerebral sac diverging into two lateral masses. The frontal lobes of the hemispheres were represented by a single mass, containing the conjoined lateral ventricles; the temporal lobes were normally developed as regards their form and position, and separated from the common frontal lobe by a distinct fissure of Sylvius; the posterior lobes were absent, there being no posterior horn of the lateral ventricle. The posterior divergence of the cerebral sac completely exposed the thalamencephalon, mesencephalon, and cerebellum.

The dura mater and pia mater were tightly adherent to the surface of the cerebrum; the cerebral falx was absent, unless it be represented by the two diverging folds of dura mater, which enveloped the diverging lateral cerebral masses. The tentorium was represented by two crescentic folds, extending out a short distance from the petrous ridge. The longi-

tudinal sinus was, of course, entirely absent; there existed, however, two venous channels originating at the point of divergence of the cerebral lobes, whilst posteriorly they followed the divergence of the lobes to empty into the lateral sinus at the position of the jugular foramen, instead of at the normal position opposite the internal occipital prominence.

The dura mater was firmly attached at a point corresponding to the crista galli, which was, however, not developed.

The pia mater followed the dura mater, and the velum was represented by a distinct fold, which did not, however, extend into the lateral ventricles.

With the exception of the fissure of Sylvius, there were no signs of fissures or convolutions, the surface of the cerebral sacs remaining entirely smooth; the cerebellum, however, exhibited faintly marked transverse fissures.

From the disorganized condition of the brain we were unable to determine completely the structure and relative position of many of its parts, but we present the following as perhaps of interest: The lateral ventricles were represented by a single cavity, of a horse-shoe shape, its limbs passing backwards into the diverging cerebral sacs; the septum lucidum, fifth ventricle, fornix, and corpus callosum were entirely absent. No corpora striata were observed, though, from the disorganized condition of the brain, it is possible that they escaped identification. The conjoined lateral ventricle communicated by a large opening, twelve millimetres in diameter, representing the foramen of Monro, with the third ventricle. The optic thalami, pineal body, and infundibulum were present, but no trace of the pituitary body was found. The mesencephalon was normally developed, the Sylvian aqueduct passing from the third to the fourth ventricle, having above it the quadrigeminal bodies and valve, and below the cerebral peduncles. Nothing abnormal was noted in connection with the cerebellum, medulla, pons, or fourth ventricle.

On removing the brain from the cranial cavity, it was found that the olfactory lobes were absent, as were also the optic tract and chiasm; the latter being represented in the cranial cavity by a single nerve, which passed through a single optic foramen. The remaining cranial nerves appeared to be normal.

A few observations were made upon the muscles of the face and head. The occipito-frontalis was well developed; the auricular muscles were normal. Rudimentary fibres were found arising from the temporal fossa, great wing, and pterygoid process of the sphenoid and from the zygomatic arch, representing from their origin and direction the temporal, masseter, and pterygoid muscles; from the absence of the lower jaw they were naturally not normally developed. The fatty cheek-pad was well formed, but was situated upon the squamous portion of the temporal bone. An orbicularis palpebrarum encircled the single eye. With regard to the remaining facial muscles, nothing definite could be determined.

The blind pouch, situated below the superior maxillary bones, described above as communicating with the exterior by a small rounded aperture, proved to be a rudimentary mouth (stomodæum), lined with mucous membrane, and presented upon the alveolar ridge of the superior maxillary bone distinct prominences which were found to be produced by the teeth pertaining to the superior maxillæ; that is, four molars and two canines, the incisors being absent. The salivary glands were not present.

A microscopic examination of the nasal trumpet revealed the following

facts: Externally it was covered with integument, which passed into the blind sac situated at the free extremity, without exhibiting any tendency to become converted into mucous membrane. Numerous sebaceous and hair follicles and rudimentary muscular fibres were found. In the interior the lateral nasal cartilages were represented by a truncated, hollow, cartilaginous cone, compressed laterally, notched at the apex, and at the base of the inferior margin showing a tendency to inversion. In the interior of this cone was a nearly circular cavity lined with mucous membrane. This cavity was separated from the blind integumental pouch by a partition about three millimetres thick. No cartilaginous or bony septum could be discovered, either in the proboscis or at its point of attachment to the frontal region.

The eyeball was exposed *in situ* by the removal of the roof of the orbit. The levator palpebræ was represented by a single well developed fan-shaped muscle, supplied by the occulo-motor nerve, arising normally from the apex of the orbit, to be inserted into the superior tarsal cartilage. On dividing this muscle, two straight muscles, lying at equal distances from the median line, were found arising from the apex of the orbit and inserted into the anterior portion of the eyeball; these represented either two superior recti or a single rectus and a superior oblique. The absence of any intermediate tendinous portion and their similarity in origin and insertion, as well as the points hereafter described with regard to the interior of the eye, incline us to the former view. A single inferior rectus and a lateral rectus on each side were found; the right lateral rectus was joined at its anterior third by a band of muscular fibres, probably representing a single inferior oblique, arising from the left inferior wall of the orbit. An ophthalmic ganglion was found situated between the right lateral rectus and the optic nerve. This, together with the relations of the inferior oblique muscle, would indicate that the single eyeball corresponded with the right, whilst the left was undeveloped.

The sclerotic was well developed; the cornea, iris, and pupil circular; a single lens of normal shape with no signs of a groove or fusion of two lenses. The vitreous humour and ciliary processes were normal, and the choroid present as a separate tunic.

The retina was detached and too friable to admit of accurate study. On removing the retina, two coalesced cribriform spots were found, as if the optic nerve had partially subdivided at this point, although outside of the eyeball not a trace of division could be detected. On cross-section of the optic nerve at its point of passage through the sclerotic, it was found that the apparent fusion of two cribriform spots was associated with an attempt at subdivision of the optic nerve at that point, two central retinal arteries being detected, which, outside the eyeball, united to a common trunk. This was the only sign present of any attempt at subdivision, or indication of fusion in the eyeball, though no study could be made of the retina.

On examining the auditory apparatus, the two external auditory canals were found to communicate directly with the pharynx, their entrance being surrounded by two tympanic bones separated by an interval of 17 mm.; there was no sign of tympanic membranes. Lying between the tympanic bone and the periotic capsule (hyo-mandibular cleft) were found the malleus, incus, and stapes of each side. The handles of the two mallets were directed downwards, forwards, and inwards, and the head of each articulated with an incus; the processus gracilis of each malleus turned inwards towards the median line, and was united by a slender splint-like bone

with its fellow of the opposite side. This probably represented an undeveloped lower jaw. The stapes and anvil were partly surrounded by an imperfect bony canal formed by the tegmentum tympani, representing an imperfect middle ear. The two processes of each incus were directed upwards, outwards, and backwards, the long process of each articulating with the stapes whilst the short process was attached to the roof of the imperfect tympanic cavity by a distinct ligament. The stapes articulated with the oval window, and was somewhat deformed. The chain of ear ossicles was, therefore, directed from within outwards instead of the normal reversed condition.

The larynx and trachea were normally developed, the two plates of the thyroid cartilages being ununited. The epiglottis and hyoid apparatus were present, but there was no trace of a tongue. The muscles below the position of the hyoid were found in their ordinary condition, but above there was an indistinct undifferentiated muscular mass which represented the undeveloped muscles of the floor of the mouth. The pharynx was attached to the base of the skull and ended in a cul-de-sac above, separated from the stomodæum by a partition 1 cm. in breadth, no communication existing between the pharynx and the nasal cavity already described.

With regard to the skull, the following may be noted: The occipital segment is normally formed, consisting of the basi- supra- and ex-occipitals. The parietal segment has a well developed basi-sphenoid, two alisphenoids or greater wings, and is completed by well developed parietals. The frontal segment is, however, somewhat modified, the presphenoid is absent, whilst the orbito-sphenoids, or lesser wings, have been displaced backwards, uniting with the basi-sphenoid posterior to the origin of the alisphenoids. They have also approached laterally towards the median line where they have coalesced, presenting in the centre a single optic foramen. The appearance of the combined orbito-sphenoids is, therefore, that of a triangular splint-like bone, having its base resting on the internal upper edges of the approximated alisphenoids, not articulating with the orbital plates of the frontal, and its apex co-ossified with the basi-sphenoid; as a result of this, there is no pituitary fossa, but instead a bridge of bone leaving a space of about 3 mm. between it and the base of the skull. As a consequence, also, of this union of the orbito-sphenoids and absence of the presphenoid, the sphenoidal foramina are represented by a single opening situated between the internal edges of the alisphenoids, and beneath the bridge-like orbito-sphenoid.

The pterygoid processes are small and not well developed.

The frontal bone is represented by a single bone in which, however, a distinct sutural line can be detected. It is greatly malformed, having only about one-half the breadth and two-thirds the normal height. No frontal prominences can be detected, the glabellum and internal angular processes are entirely absent, the supra-orbital ridges of the two sides meeting in the middle line form a single ridge bounding the orbital cavity above. The external angular processes are well developed, and articulate with the malar bones below. There is a single orbital plate with a large deficiency in the centre corresponding to the ethmoidal notch: posteriorly it articulates with the alisphenoids instead of the orbito-sphenoids. The ethmoids are entirely absent. The temporal bone has all of its elements represented, the petromastoid presenting its normal appearance and relations, with the exception that a thin plate of bone has been

given off inferiorly to insheathe an imperfect tympanic cavity, taking the place, to a certain extent, of the tympanic bones which we have described above as being displaced to an abnormal position at the base of the skull. The squamosal element has likewise been displaced inwardly so that at the base of the skull they are but five millimetres apart instead of five centimetres, which is about the space normally separating them in a fœtal skull of the same age. There is also no trace of a glenoid cavity or fissure, unless the latter be represented by a wide gap lying between the squamosals and the thin plate of bone already described as given off by the petromastoids.

The face is in an exceedingly rudimentary condition, but few of its bones being developed. The ento-pterygoids are present, but have not united with the pterygoid process of the sphenoid, being, indeed, situated posteriorly to them. The palate bones are likewise feebly developed, being represented by two small plates of bone lying between the pterygoid processess. Of the two superior maxillaries, only the bodies are developed, and of these, the alveolar borders form the greater part, the palate plates, parts bounding the nasal cavities, tuberosity, infra-orbital ridges, and premaxillary portion and nasal spines being entirely absent. A small orbital surface, formed by the conjoined plates of the two bones, exists, however, and the two infra-orbital ridges are well marked.

The malar bones are normally formed, but owing to the arrested development of the superior maxillary bone, they approach one another and are separated in the median line by a distance of five millimetres instead of four centimetres, the normal intermalar space. On this account they form almost entirely the inferior and external boundaries of the orbit. A distinct zygomatic process is present, but it does not unite with the zygomatic process of the squamosal.

Of the remaining bones of the face, the lachrymals, nasals, turbinates, and vomer, are entirely absent, whilst the inferior maxillary is only represented by the small splint-like bone previously described.

The single orbit is formed by the following bones; Above by the orbital plate of the frontal, posteriorly by the orbital surfaces of the alisphenoides, inferiorly and laterally by the malars and conjoined orbital plates of the superior maxillaries. The elements absent that enter into the normal construction of the orbit are the orbital surfaces of the ethmoid, the lachrymal, orbito-sphenoids, and orbital plates of the palate bone. The shape of the orbit does not differ widely from the normal form, and has entering into it the following apertures: A single optic foramen, a single sphenoidal fissure formed by the union of the right and left, two rotund foramina, and two spheno-maxillary fissures.

The nasal cavities are entirely absent.

The chief interest to be derived from the study of monstrosities, such as described above, lies in the explanation of the causes which have led to the various deviations from their normal development. While we have found accounts of numerous monsters which, from their general appearance, probably resembled, in many points, the one described, the narrow-mindedness of custodians of museums, who are satisfied with a general description of the exterior and are then content to suspend the specimens in jars for the amazement and *instruction* of the curious, has greatly interfered with a scientific knowledge of the laws of teratology. Without a

thorough dissection of the internal parts nothing, as to the causes producing deformity, can be determined.

A study of the monstrosity described above shows that all the deviations from the normal type can be explained by the modification or non-development of certain parts.

Normally, the brain develops in the following manner: The anterior end of the primitive medullary tube dilates into three cerebral vesicles; of these the first remains as the thalamencephalon, sending off anteriorly two prosencephalic buds to form the hemispheres. These, in their turn, send off each a secondary bud, the rhinencephalon, or olfactory lobe. In the case before us, however, the thalamencephalon has been normally developed, but instead of sending off a pair of prosencephalic buds, but a single bud was formed, which, however, partially divided posteriorly into two. The reason for considering that but a single bud was given off, rather than that fusion occurred between two primitive buds, lies in the absence of any partition wall between the homologues of the two lateral ventricles. No secondary buds, or rhinencephala, were given off, hence the entire absence of olfactory lobes.

In the same manner, we believe but a single optic bud appeared; hence the median position of a single eye.

The explanation ordinarily given for the approximation of two eyes, or the presence of a single eye in the comparatively well-described group of simple cyclops monstrosities, is that two primitive optic buds have converged and coalesced in the median line; in many cases this may be the correct explanation; but where, as in the present instance, but a single eye has appeared, or where there is a close coalescence of two eyes, it appears to us much more probable that but a single median primitive optic bud has been given off from the thalamencephalon, and that this bud either remains entirely single, thus producing but a single eye as in this case, or that in those cases which present an apparent fusion of two eyeballs and optic tracts, the primitive median optic bud has subdivided more or less completely into two. It is difficult to conceive that after the coalescence of two primitively distinct optic buds, the mesoblastic tissues, which go to form the tissues of the eyeball, should be able so to adjust themselves as to produce a single perfect and normally developed eye.

Owing to this development of a single optic bud, or it may be the convergence and coalescence of two primitively distinct buds, the molecular arrangement of the embryonal cells situated at the base of the skull is so disturbed that we find in all these cases of cyclops that the fronto-nasal process fails to develop. As a result of this follows the entire absence of all the bony parts, developing normally from this process, viz., the ethmoid, nasals, lachrymals, vomer, and pre-maxillary bone, and hence the presence of the eye beneath a nasal proboscis belonging only to the skin.

The absence of the lower jaw and malformation of the auditory apparatus, an arrangement which, so far as we have been able to learn, is entirely unique, may be explained as depending upon defective development of the first visceral arch. Normally from the base of the primitive cartilaginous cranium we have given off on each side two cartilaginous ventral rods, which pass down into the anterior visceral arches. From the first of these we have given off the palato-pterygoid plate, which passes out into the maxillary process, to form the basis of the upper jaw, whilst the continuation of the rod extends downwards, under the name of the cartilage of Meckel, into the mandibular arch and forms the basis of the lower jaw. The second cartilaginous rod forms the hyoid series. In the higher vertebrates the proximal element of the mandibular arch is converted into the malleus, the homologue of the quadrate bone of lower forms, whilst the proximal element of the hyoid arch is converted into the incus, the homologue of the hyo-mandibular. The remaining auditory ossicle or stapes is formed, according to most authorities, from a part of the periotic capsule.

In the present case, the deviations from the normal form have been produced by the irregular development of the cartilages of Meckel. These, instead of passing downwards, turned inwards towards the base of the skull, to meet in the median line. Hence, therefore, the lower jaw, which consists of a pair of membrane bones developed in the tissues surrounding the cartilages of Meckel, is represented by a small, slender, splint-like bone, lying at the base of the skull, at a point corresponding in position with that attributed above to the irregularly developed cartilages of Meckel. From this arises also the displaced position of the ear ossicles, which occupy a position at the base of the skull corresponding closely to their normal position in an early stage of development of the hyomandibular cleft.

The hyomandibular cleft, or space between the primitive mandibular and hyoid visceral arches, remains in the higher vertebrates as the external auditory meatus, tympanic cavity, and Eustachian tube. Owing to the approximation of all the parts towards the median line, this canal has been in this specimen very much shortened; the tympanic cavity exists, as we have already described, in an imperfect condition, bounded by the petro-mastoid and squamosal elements of the temporal bone alone, the tympanic bone, from its displaced condition, not entering into its construction. As a result of this, we find that the portion of the canal corresponding to the external auditory meatus is quite short, and enters the pharynx below the position of the middle ear, though communicating freely with it. A Eustachian tube does not exist, since the ear ossicles are not entirely closed within the petro-mastoid bone, the malleus lying entirely within the cavity of the pharynx. The internal ear was normal.

Case I. *Cyclopian Monster.*[1]—Dr. D. W. Richardson was called, on the evening of Dec. 7, 1881, to attend Mrs. D., a multipara, in labour, but did not reach the patient until the child was born. According to the father's statement the child lived ten or fifteen minutes after birth. On inquiry, the doctor could not find anything abnormal in the labour, only an excessive amount of the liquor amnii.

The next morning he came in possession of the child; and brought it to my office for examination.

It was a female, born at term, weighed four pounds, and body well developed. It had but one eye, of normal size, situated in the middle of its face, and a little above where the mouth should have been.

The nose consisted of a mere conical fleshy semi-tube, starting out by its smaller end, immediately above the eye; its dimensions being a little over an inch in length, and about half an inch in diameter at its free extremity. It contained no cartilage, and, instead of two anterior nares, there was but one small, round opening, or canal, about one-fourth of an inch in depth. It was quite soft and flexible, and hung loosely over the eye.

The maxillary bones were united, and no line between them could be determined. The mouth consisted of a small aperture, and situated under a small fold of skin, immediately beneath the symphysis of the inferior maxillary bone. A probe could be passed through this small opening into the throat.

Fig. 1.

Cyclopian monster. Head flexed.

Fig. 2.

Cyclopian monster. Head extended.

The ears were of normal size, but situated much lower, and approaching each other.

There were apparently no external auditory canals, but, instead, there were two small depressions.

[1] Reported by M. A. Koogler, M.D., in the American Journal of the Medical Sciences, for July, 1882.

The ears were symmetrical, and the lobes and the cartilages below the tragi and anti-tragi were absent, and in their place, running inwards and approaching one another, were two slit-like openings, about one-third of an inch in length. These openings communicated with the larynx, and a probe could be passed in the one and out of the other. The father stated that during the time it lived, "its breathing was through these openings." The appearances of the monster are well shown in the accompanying drawings (Figs. 1 and 2).